Short reports

Cardiac arrhythmias misdiagnosed as epilepsy

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SUMMARY A mother and three children presenting with syncope induced by exercise and emotion were diagnosed as epileptic. They, and three symptom free children, showed frequent ventricular and supraventricular tachyarrhythmias on ambulatory electrocardiographic monitoring. Three died before the correct diagnosis of disordered sympathetic innervation of the heart was made, but episodes of syncope and cardiac arrhythmias in the survivors have been successfully treated by propranolol.

It is well known that episodic loss of consciousness may have a cardiac rather than cerebral origin. We present a family where these episodes were misdiagnosed and treated as epilepsy.

Case reports

A mother and three of her thirteen children (Fig. 1) presented with syncopal attacks which had strikingly similar features. In each case the patient would fall to the ground unconscious without warning, no tonic or clonic movements occurred, and recovery of consciousness took place within a few minutes. The episodes had their onset in late childhood and occurred only on exercise (for example running, walking up hill, lifting a heavy load) or with emotion (for example fear, anger).

The mother (patient II.7 in Fig. 1) developed syncope on exertion or when emotionally aroused at the age of 11 years, attacks occurring about once a

month. No other family member of her or her parents' generation suffered these attacks. Epilepsy was diagnosed in adolescence and a variety of anticonvulsant drugs were tried, without success. An electroencephalogram was normal. The attacks continued into adult life until the age of 46 years, when it was noted during one of her episodes that her heart was not beating. When assessed in a cardiac clinic she had no physical abnormalities, and a chest radiograph and echocardiogram were normal. An electrocardiogram was normal, with a sinus rhythm of 60 per minute and a QT interval corrected for heart rate (QT_c) of 0.36 seconds (normal range 0.32 to 0.46 seconds). An ambulatory electrocardiogram recording was grossly abnormal showing episodes of severe ventricular arrhythmias even in the absence of symptoms. These included frequent multifocal ventricular ectopics with bigeminy, couplets, triplets, and short runs of ventricular tachycardia. Occasional brief spells of supraventricular tachycardia were also seen. The presence of periods of sinus bradycardia (slowest heart rate 40 per minute) led to a diagnosis of sinus node disease ('tachy-brady' syndrome), and since it was felt that episodes of asystole were responsible for her symptoms, a transvenous pacemaker was inserted. Her syncopal episodes were perhaps less frequent after this, but more severe. A year later she collapsed and was resuscitated with great difficulty—she never regained consciousness and died a few weeks later. The pacemaker was found to be functioning correctly.

Three children had similar episodes, starting in late childhood and precipitated by exercise and emotion. Patient III.1 was a tall, thin youth who attended a school for the educationally subnormal (M). He was diagnosed as an epileptic and treated unsuccessfully with phenobarbitone. An electroencephalogram and electrocardiogram (heart rate 55 per minute, QT_c 0.37 seconds) were normal. He died suddenly at the age of 19 while playing football. A brother (patient III.3) with identical episodes was also labelled epileptic but not investi-

Fig. 1 Family tree (with conventional pedigree notation).

gated or treated. He collapsed and died at the age of 16 while carrying a sack of potatoes.

Patient III.8 is a thin, 14 year old boy with no physical abnormalities who attends a school for the educationally subnormal (M). His syncopal attacks started at the age of 11 years; he was diagnosed as epileptic and treated unsuccessfully with phenobarbitone. An electroencephalogram and electrocardiogram (rate 60 per minute, QT_c 0.42 seconds) were normal. Further investigation was carried out when it was learned that his mother had cardiac arrhythmias. A chest radiograph and echocardiogram were normal, but ambulatory electrocardiographic monitoring showed arrhythmias similar to his mother's (Fig. 2). The slowest heart rate was 55 per minute. An exercise test had to be stopped after only 45 seconds as he became distressed and frightened and his electrocardiogram showed very frequent ventricular ectopics with short runs of ventricular tachycardia. As with his mother, a pacemaker was inserted because he was thought to

have sinus node disease, but his syncopal attacks and arrhythmias continued. After a few months, a diagnosis of disordered sympathetic innervation of the heart was made (similar to the prolonged QT syndrome but with a normal QT interval) and he was started on regular propranolol. On a daily dose of 160 mg, he has been free from further attacks for 18 months and repeated 24 hour ambulatory electrocardiographic recordings have been normal with no ventricular or supraventricular arrhythmias.

The other 10 siblings were investigated by chest radiograph, electrocardiogram, echocardiogram, and 24 hour ambulatory monitoring. It was discovered that three younger children (III.10, III.11, and III.12) age 7, 8, and 10 years showed identical arrhythmias to those of their mother and affected brothers. They have no symptoms, attend normal schools, and show no physical abnormalities apart from being rather thin. All other investigations were normal (heart rate 96, 96, and 90 per minute; QT_c 0.46, 0.41, and 0.39 second respectively). These three children have been treated with propranolol (60 mg/day), they remain symptom free, and repeated 24 hour ambulatory electrocardiographic monitoring shows complete abolition of the ventricular and supraventricular arrhythmias.

Discussion

It seems likely that this family suffer from disordered sympathetic innervation of the heart, a variant of the prolonged QT syndrome.¹ Structural heart disease such as cardiomyopathy or a prolapsing mitral valve has been largely excluded by examination and cardiac investigation. Disordered generation of the cardiac impulse (sinus node disease) was the initial diagnosis, although this is rare in children except after cardiac surgery.² The diagnosis was made because of periods of sinus bradycardia, but these were not severe and the symptoms were unaffected by the insertion of a demand pacemaker. The prolonged QT syndrome may occur alone³ as an autosomal dominant condition or with severe nerve deafness⁴ as a recessive disorder. A variant has recently been described and reviewed^{5,6} in which the QT interval is normal but where severe ventricular arrhythmias occur on exercise or emotion. Syncope is probably due to ventricular fibrillation induced by the R on T phenomenon. It is not certain whether the normal QT patients are a distinct subgroup physiologically and genetically or whether they represent a 'forme fruste' of the prolonged QT syndrome. Beta blockade is the first treatment in all variants and may be life saving. Left cervical sympathetic ganglionectomy has been used in resistant cases.

Fig. 2 Excerpts from a 24 hour ambulatory electrocardiographic recording of the 14 year old boy with syncopal attacks (patient III.8). He was symptom free during the period of the recording which shows:

(a) sinus rhythm, (b) ventricular ectopics and a brief run of ventricular tachycardia, (c) irregular ventricular tachycardia, (d) possible supraventricular tachycardia.

Syncope on exertion or emotion, whether or not accompanied by convulsions, is rarely epileptic and should be investigated for a cardiac cause. Initial investigations should include a chest radiograph, echocardiogram, and standard electrocardiogram; these should be followed by an examination of the heart rate and rhythm during exercise. This can be done by means of an exercise test or, more simply in children, by 24 hour ambulatory electrocardiographic monitoring. An event marker on the monitor can be pressed if the child develops any symptoms during the 24 hours of the recording, so that the heart rate and rhythm at the time of the event can be identified when the tape is being examined. If these more extended tests of heart rhythm are not performed, this particular cardiac rhythm disorder, which is dangerous but treatable, will be missed.

We thank Dr D I Johnston, Dr G K Morris, and Dr V Redding for

their help with this family. Dr Southall is senior research fellow of the British Heart Foundation.

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Received 18 July 1984

Perinatal hepatitis B virus detection by hepatitis B virus-DNA analysis

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SUMMARY Maternal transmission of hepatitis B virus infection in relation to the hepatitis B e antigen/antibody system and serum hepatitis B virus-DNA were evaluated. Results indicate that hepatitis B virus-DNA analysis can identify hepatitis B serum antigen positive mothers who may transmit infection to their offspring.

Perinatal transmission of hepatitis B virus infection from hepatitis B surface antigen positive mothers to their infants seems to be associated primarily with the presence of maternal hepatitis B e antigen.¹ Most mothers who are hepatitis B e antigen positive transmit the infection to their infants, whereas those who have antibody to hepatitis e antigen or are negative for both hepatitis B e antigen and antibody rarely infect their infants.² It has recently been shown, however, that perinatal transmission of hepatitis B virus infection may occur frequently, even in mothers positive for hepatitis B e antibody

or negative for both hepatitis B e antigen and antibody.^{3,4} We evaluated the risk of perinatal hepatitis B virus transmission from asymptomatic hepatitis B surface antigen positive mothers in relation to the hepatitis B e antigen/antibody system and serum hepatitis B virus-DNA.

Patients and methods

While screening for hepatitis B surface antigen in the antenatal clinic, we identified 96 hepatitis B surface antigen positive pregnant women out of 1500 tested. All were asymptomatic. Plasma from these women was stored frozen at -70°C until assayed for the following: hepatitis B core antibody, hepatitis B surface antibody, and hepatitis B e antigen and antibody (Abbott Laboratories); liver function tests (measured by standard laboratory methods); and hepatitis B virus-DNA (spot technique according to Scotto *et al.*, 1983).⁵ These determinations were also carried out in the respective offspring at birth and, thereafter, monthly for one year. All babies of